Sézary syndrome

Sézary syndrome is an aggressive form of a type of blood cancer called cutaneous T-cell lymphoma. Cutaneous T-cell lymphomas occur when certain white blood cells, called T cells, become cancerous; these cancers characteristically affect the skin, causing different types of skin lesions. In Sézary syndrome, the cancerous T cells are called Sézary cells and are found in the skin, lymph nodes, and blood. A characteristic of Sézary cells is an abnormally shaped nucleus, described as cerebriform.

People with Sézary syndrome develop a red, severely itchy rash (erythroderma) that covers large portions of their body. Sézary cells are found in the rash. However, the skin cells themselves are not cancerous; the skin problems result when Sézary cells move from the blood into the skin. People with Sézary syndrome also have enlarged lymph nodes (lymphadenopathy). Other common signs and symptoms of this condition include hair loss (alopecia), thickened skin on the palms of the hands and soles of the feet (palmoplantar keratoderma), abnormalities of the fingernails and toenails, and lower eyelids that turn outward (ectropion). Some people with Sézary syndrome are less able to control their body temperature than people without the condition.

The cancerous T cells can spread to other organs in the body, including the lymph nodes, liver, spleen, and bone marrow. In addition, affected individuals have an increased risk of developing another lymphoma or other type of cancer.

Sézary syndrome occurs in adults over age 60 and progresses rapidly; historically, affected individuals survived an average of 2 to 4 years after development of the condition, although survival has improved with newer treatments.

Although Sézary syndrome is sometimes referred to as a variant of another cutaneous T-cell lymphoma called mycosis fungoides, these two cancers are generally considered separate conditions.

Frequency

Sézary syndrome is a rare condition, although its prevalence is unknown. It is the second most common form of cutaneous T-cell lymphoma after mycosis fungoides, accounting for approximately 3 to 5 percent of cases of cutaneous T-cell lymphoma.

Genetic Changes

The cause of Sézary syndrome is unknown. Most affected individuals have one or more chromosomal abnormalities, such as the loss or gain of genetic material. These abnormalities occur during a person's lifetime and are found only in the DNA of cancerous cells. Abnormalities have been found on most chromosomes, but some

regions are more commonly affected than others. People with this condition tend to have losses of DNA from regions of chromosomes 10 and 17 or additions of DNA to regions of chromosomes 8 and 17. It is unclear whether these alterations play a role in Sézary syndrome, although the tendency to acquire chromosomal abnormalities (chromosomal instability) is a feature of many cancers. It can lead to genetic changes that allow cells to grow and divide uncontrollably.

Inheritance Pattern

The inheritance pattern of Sézary syndrome has not been determined. This condition occurs in people with no history of the disorder in their family and is not thought to be inherited in most cases.

Other Names for This Condition

- Sezary erythroderma
- Sezary syndrome
- Sezary's lymphoma

Diagnosis & Management

Genetic Testing

- Genetic Testing Registry: Sezary syndrome
  https://www.ncbi.nlm.nih.gov/gtr/conditions/C0036920/

Other Diagnosis and Management Resources

- Cancer Research UK: Treatments for Cutaneous T-Cell Lymphoma
  http://www.cancerresearchuk.org/about-cancer/type/non-hodgkins-lymphoma/about/types/cutaneous-t-cell-lymphoma#treat
- Cutaneous Lymphoma Foundation: Sezary Syndrome Fast Facts
  http://www.clfoundation.org/sites/default/files/content/Fact%20Sheet_SS.pdf
- Lymphoma Research Foundation: Cutaneous T-Cell Lymphoma Treatment Options
  http://www.lymphoma.org/site/pp.asp?c=bkLTKaOQLmK8E&b=6300151#TreatmentsOptions
- National Cancer Institute: Mycosis Fungoides and the Sézary Syndrome Treatment
  https://www.cancer.gov/types/lymphoma/patient/mycosis-fungoides-treatment-pdq
- National Organization for Rare Disorders (NORD) Physician Guide
  http://nordphysicianguides.org/cutaneous-t-cell-lymphoma-ctcl/

General Information from MedlinePlus

- Diagnostic Tests
  https://medlineplus.gov/diagnostictests.html
- Drug Therapy
  https://medlineplus.gov/drugtherapy.html
- Genetic Counseling
  https://medlineplus.gov/geneticcounseling.html
- Palliative Care
  https://medlineplus.gov/palliativecare.html
- Surgery and Rehabilitation
  https://medlineplus.gov/surgeryandrehabilitation.html

Additional Information & Resources

MedlinePlus

- Health Topic: Cancer--Living with Cancer
  https://medlineplus.gov/cancerlivingwithcancer.html
- Health Topic: Lymphoma
  https://medlineplus.gov/lymphoma.html

Genetic and Rare Diseases Information Center

- Sezary syndrome
  https://rarediseases.info.nih.gov/diseases/7629/sezary-syndrome

Additional NIH Resources

- National Cancer Institute: Mycosis Fungoides and the Sézary Syndrome Treatment
  https://www.cancer.gov/types/lymphoma/patient/mycosis-fungoides-treatment-pdq

Educational Resources

- Cutaneous Lymphoma Foundation: Sezary Syndrome Fast Facts
  http://www.clfoundation.org/sites/default/files/content/Fact%20Sheet_SS.pdf
- Disease InfoSearch: Sezary syndrome
  http://www.diseaseinfosearch.org/Sezary+syndrome/6534
- Leukemia and Lymphoma Society: Cutaneous T-Cell Lymphoma Facts
  http://www.lls.org/sites/default/files/file_assets/FS5_Cutaneous%20T-Cell%20Lymphoma_2014_Final.pdf
- Lymphoma Research Foundation: Cutaneous T-Cell Lymphoma
  http://www.lymphoma.org/atf/cf/%7BAAF3B4E5-2C43-404C-AFE5-FD903C87B254%7D/LRF_FACTSHEET_CTCL_2013.PDF

- MalaCards: sezary's disease
  http://www.malacards.org/card/sezarys_disease
- Orphanet: Sézary syndrome
  http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=3162

Patient Support and Advocacy Resources

- American Cancer Society
  https://www.cancer.org/
- Cutaneous Lymphoma Foundation
  http://www.clfoundation.org/
- Leukemia and Lymphoma Society
  http://www.lls.org/
- Leukemia and Lymphoma Society: Co-Pay Assistance Program
  http://www.lls.org/support/financial-support/co-pay-assistance-program
- Lymphoma Research Foundation
  http://www.lymphoma.org/
- National Organization for Rare Disorders (NORD): Cutaneous T-Cell Lymphomas
  https://rarediseases.org/rare-diseases/cutaneous-t-cell-lymphomas/

ClinicalTrials.gov

- ClinicalTrials.gov
  https://clinicaltrials.gov/ct2/results?cond=%22Sezary+syndrome%22

Scientific Articles on PubMed

- PubMed
  https://www.ncbi.nlm.nih.gov/pubmed?term=%28Sezary+Syndrome%5BMAJR%5D%29+AND+%28Sezary+syndrome%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+720+days%22%5Bdp%5D

Sources for This Summary

- Campbell JJ, Clark RA, Watanabe R, Kupper TS. Sezary syndrome and mycosis fungoides arise from distinct T-cell subsets: a biologic rationale for their distinct clinical behaviors. Blood. 2010 Aug 5;116(5):767-71. doi: 10.1182/blood-2009-11-251926. Epub 2010 May 18.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/20484084
  Free article on PubMed Central: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2918332/
- Caprini E, Cristofoletti C, Arcelli D, Fadda P, Citterich MH, Sampogna F, Magrelli A, Censi F, Torreri P, Frontani M, Scala E, Picchio MC, Temperani P, Monopoli A, Lombardo GA, Taruscio D, Narducci MG, Russo G. Identification of key regions and genes important in the pathogenesis of sezary syndrome by combining genomic and expression microarrays. Cancer Res. 2009 Nov 1;69(21): 8438-46. doi: 10.1158/0008-5472.CAN-09-2367. Epub 2009 Oct 20.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/19843862

- Hwang ST, Janik JE, Jaffe ES, Wilson WH. Mycosis fungoides and Sézary syndrome. Lancet. 2008 Mar 15;371(9616):945-57. doi: 10.1016/S0140-6736(08)60420-1. Review.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/18342689
- Izykowska K, Przybylski GK. Genetic alterations in Sezary syndrome. Leuk Lymphoma. 2011 May; 52(5):745-53. doi: 10.3109/10428194.2010.551159. Epub 2011 Feb 16. Review.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/21323514
- Laharanne E, Oumouhou N, Bonnet F, Carlotti M, Gentil C, Chevret E, Jouary T, Longy M, Vergier B, Beylot-Barry M, Merlio JP. Genome-wide analysis of cutaneous T-cell lymphomas identifies three clinically relevant classes. J Invest Dermatol. 2010 Jun;130(6):1707-18. doi: 10.1038/jid.2010.8. Epub 2010 Feb 4.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/20130593
- Rosen ST, Querfeld C. Primary cutaneous T-cell lymphomas. Hematology Am Soc Hematol Educ Program. 2006:323-30, 513. Review.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/17124079
- Wong HK, Mishra A, Hake T, Porcu P. Evolving insights in the pathogenesis and therapy of cutaneous T-cell lymphoma (mycosis fungoides and Sezary syndrome). Br J Haematol. 2011 Oct; 155(2):150-66. doi: 10.1111/j.1365-2141.2011.08852.x. Epub 2011 Aug 25. Review.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/21883142
  Free article on PubMed Central: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4309373/
- Yamashita T, Abbade LP, Marques ME, Marques SA. Mycosis fungoides and Sézary syndrome: clinical, histopathological and immunohistochemical review and update. An Bras Dermatol. 2012 Nov-Dec;87(6):817-28; quiz 829-30. Review.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/23197199
  Free article on PubMed Central: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3699909/
- van Doorn R, van Kester MS, Dijkman R, Vermeer MH, Mulder AA, Szuhai K, Knijnenburg J, Boer JM, Willemze R, Tensen CP. Oncogenomic analysis of mycosis fungoides reveals major differences with Sezary syndrome. Blood. 2009 Jan 1;113(1):127-36. doi: 10.1182/blood-2008-04-153031. Epub 2008 Oct 1.
  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/18832135

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